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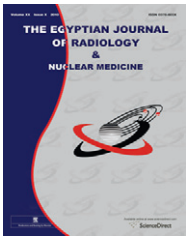

Intrahepatic vascular shunts: Strategy for early diagnosis, evaluation and management

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KEYWORDS

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Management

Abstract *Purpose:* To propose a strategy for early diagnosis and management of intrahepatic vascular shunts (IHVSs).

Patients and Methods: We systematically screened 3143 patients liable to develop IHVSs. Diagnosed patients were comprehensively evaluated using color Doppler and multiphasic CT. Our management strategy based on imaging and clinical findings and categorized patients into three groups with different lines of management and we assessed the degree of shunt occlusion and clinical outcome.

Results: IHVSs were diagnosed in 134 patients. Two types of shunts were identified; arteriovenous (72%) and venovenous (28%). Small simple shunts (32%) showed no morphological or hemodynamic changes. Small complex shunts (57%) showed mild alterations in Doppler spectra. Large and aneurismal shunts (11%) showed increased diameters and flow velocities of the shunt vessels with alteration of flow pattern. By CT the vessels opacified during the early arterial phases in

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arteriovenous, and during the portovenous phase in venovenous shunts. During management, group I included 42%; 38% were conservatively managed and 4% underwent TIPS. Group II included 57% who underwent successful embolization in 50%, and <80% reduction in 7% of patients who underwent surgery. Direct surgery was selected for 1% of patients.

Conclusion: Using color Doppler ultrasonography complemented by multiphasic CT allowed early diagnosis and evaluation of IHVSs and selection of the suitable treatment. Our management strategy allowed practical therapeutic implications and yielded good results.

1. Introduction

Intrahepatic vascular shunts (IHVSs) are abnormal communications between intrahepatic vasculature involving the arterial, portal or hepatic venous systems [1,2]. The etiology of these shunts is controversial and may be either acquired, as those associated with cirrhosis and/or hepatocellular carcinoma, those that occur after traumatic injuries to the liver or interventional transhepatic procedures (including liver biopsy, transhepatic cholangiography, or biliary surgery), or they may appear in the form of congenital and idiopathic vascular malformations, as in Rendu-Osler-Weber syndrome (hereditary hemorrhagic telangiectasia) [3–5].

The recognition of intrahepatic vascular shunts by clinical criteria alone has long been considered uncommon. They are probably underestimated regarding their clinical relevance and frequency of appearance. Congestive cardiac failure, portal hypertension, portosystemic encephalopathy, cholangitis, and atypical cirrhosis have been reported as possible serious complications related to this condition. Thus, a correct diagnosis is important, and diagnostic imaging has a fundamental role in identification and evaluation of shunts and determination of appropriate management [4,6,7].

Ultrasonography, especially in association with color Doppler has proved useful in the detection of intrahepatic vascular shunts and has the additional advantage over other imaging modalities that it enables recognition of flow direction, flow velocity, and type of blood flow (i.e., arterial, portal, or hepatic venous) noninvasively [2,3,6]. Those are important diagnostic aspects for the different types of vascular malformations. The introduction of multidetector CT, particularly in association with a multiphasic study protocol, and angiographic reconstructions makes their identification easier than in previous decades [4,8].

The purpose of the present study is to propose a strategy for early diagnosis, and management of intrahepatic vascular shunts based on our experience and a review of the literature.

2. Patients and methods

This prospective study was conducted between April 2008 and October 2010 on 3153 consecutive patients who were liable to develop intrahepatic vascular shunts (IHVSs). Ten patients; six had renal insufficiency and four had history of severe adverse reactions to iodinated contrast agents were primarily excluded from the study, so the total number of patients included was 3143 patients. We proposed our systematic screening program as a part of follow up of patients who had diagnosed liver cirrhosis (n = 2318) and HCC (n = 752) based on US and triphasic CT findings, 916 of them had previously underwent one or more interventional procedures including radiofrequency ablation, liver biopsy and/or and percutaneous ethanol injection. Our study also included patients who had traumatic liver injury (n = 29), patients who underwent transhepatic biliary drainage (n = 21), patients with positive family history of hereditary hemorrhagic telangiectasia (HHT) (n = 15) and patients with Budd-Chiari syndrome (n = 8). All the patients were subjected to detailed history taking and full clinical examination prior to imaging evaluation. Our evaluation of intrahepatic vascular shunts involved two levels of diagnostic examinations. The first was gray scale ultrasound completed by color and power-Doppler imaging and the second was multiphasic CT study. Targeted digital subtraction angiography (DSA) was performed as recommended by our team in the next step of the diagnostic ladder whenever intervention was indicated. The gold standard of this study was based on the information obtained from combined imaging findings and/or surgery.

2.1. Imaging studies

2.1.1. Ultrasonography

We used sonographic machines with real time B-mode imaging, coupled with pulsed, color and power Doppler, with 3.5 and 5 MHz convex transducers (Siemens sienna Erlangen, Germany, Nemio XG and Xario, Toshiba, Tokyo, Japan). The sonographic studies began with a preliminary morphologic evaluation of the liver and abdomen with special emphasis on the intrahepatic vasculature to screen for the presence and identity of a vascular shunt, and to evaluate the hemodynamic effects of the shunt on intrahepatic circulation.

2.1.2. Computed tomography

Computed tomography of the abdomen was performed for patients in whom intrahepatic vascular shunts were identified by ultrasound to confirm ultrasound findings and assess intrahepatic parenchymal perfusion changes. We used 16-detector CT scanners (Toshiba Aquillion, Tokyo, Japan and Siemens Somatom, definition AS, Germany). Gastrointestinal contrast medium was not administered. CT scanning started with an unenhanced CT acquisition then intravenous bolus injection of nonionic contrast material (Omnipaque, iohexol 350 mg/mL, Amersham Health) was administered at a dose of 1.5–2 mL/kg body weight with a maximal amount of 150 mL and a flow rate of 4 mL/s through a 22-gauge IV catheter inserted into an antecubital vein using an automatic injector (MEDRAD). Four sets of post contrast images were acquired in a craniocaudal direction after injection of the contrast medium. The scanning delay was individualized for each patient by using bolus-tracking software that measures the inflow of

contrast material in a region of interest placed on aorta at the level examined and automatically triggers the start of the examination when it registers a density of 100 HU. The first and second acquisitions were used for early and delayed hepatic arterial phase imaging, the third acquisition for portal venous phase imaging, and the fourth acquisition to image the hepatic venous phase.

Multiphasic CT scans were acquired, starting at the hepatic dome and proceeding in a caudal direction to the lower margin of the kidneys. Images were obtained during a single breath hold. The following parameters were used: section collimation, 0.75 mm; section thickness, 1 mm; table feed, 12 mm per rotation; rotation time, 0.5 s per rotation; 160 mAs; and 120 kVp. Images were reconstructed at an interval of 0.7 mm. The acquired data were transferred to a workstation equipped with a dedicated 2D and 3D reconstruction softwares (Vitrea 2.2 and Syngo fast view) which allowed processing of multiplanar reformatting (MPR), three-dimensional maximum intensity projection (MIP), and volume rendering (VR) programs, to generate multiplanar angiographic reconstructions.

2.2. Imaging evaluation and analysis

Imaging diagnosis of a shunt relied on direct detection of an abnormal intrahepatic cystic or tubular structure that communicates two vessels. For further evaluation of a shunt the following parameters were analyzed by each modality; type (determined from recognition of the identity of inflow and outflow vessels), size (small, large or aneurismal), angioarchitecture (simple or complex) and distribution (localized or diffuse). The diameters of inflow and outflow vessels were also assessed. Simple shunt has a single inflow vessel connected to a single outflow vessel via a single communication. Complex shunt consists of a network of inflow, outflow and communicating vessels [7]. Any other associated intra or extrahepatic findings were also assessed.

Furthermore, Doppler tracing was performed to assess the inflow and outflow vessels both quantitatively (peak systolic and diastolic flow velocities and resistivity index in hepatic artery, mean flow velocity in portal vein, diastolic peak flow velocity in hepatic vein) and qualitatively (flow pattern and flow direction). Quantification of blood flow volume in the shunt was measured by multiplying the lumen area by the mean velocity. The shunt ratio was calculated by dividing the total blood flow volume in the shunt by that in the inflow vessel [7,9,10].

Diagnosis and differentiation of various types of shunts on CT relied on other additional indirect signs reflecting the blood flow alteration induced by the shunt. We focused on analyzing the phase of contrast at which the inflow and outflow vessels and their communicating shunt vessels were opacified and whether there was any associated areas of transient hepatic attenuation differences (THADs) reflecting underlying intraparenchymal perfusion disorders.

2.3. Strategy used for shunt management

In order to select appropriate management for patients diagnosed as having IHVSs, our team proposed a practical strategy with therapeutic implication based on both imaging and clinical findings and categorized patients into three groups as follows:

Group I included asymptomatic patients with small non neoplastic shunts and with shunt ratio $<30\%$. They were followed up by duplex Doppler and clinical examination at 3–6 months interval. If the shunt did not change or regressed and no signs or symptoms developed, then conservative management was applied according to the type of shunt. If the shunt increased in size or new shunts developed, shunt ratios exceeded 30% or patients developed signs and symptoms, then, interventional treatment was applied to prevent long term development of complications or to improve the patients' clinical condition.

Group II included patients with symptomatic shunts, patients with large or aneurismal shunts, patients with shunt ratio $>30\%$ or patients with neoplastic shunts. They directly underwent interventional management for their shunts alone or as a bridge for surgery depending on the results of the interventional procedure applied.

Group III included patients with diffuse shunts or patients with shunt ratio $>60\%$. They were planned directly for surgery.

2.4. Interventional procedures

All angiographic examinations were performed using digital subtraction angiography unit (Infinex, Toshiba Japan). Transcatheter embolization was performed for patients with arteriovenous or portosystemic shunts via one of the following access routes depending on the type of shunt:

- In patients with arteriovenous shunts a femoral artery approach was adopted, using the Seldinger technique under local anesthesia. Baseline digital subtraction angiography of the celiac trunk and the superior mesenteric artery was performed to visualize the liver vasculature and evaluate the portal vein patency. The common hepatic artery, right and left hepatic arteries were cannulated, and more superselective cannulation of the feeders to the arteriovenous shunts was performed, if indicated, with or without the use of microcatheters (Terumo progreat 2.7 F, Terumo, Japan). To avoid passage of the embolic agent into the systemic circulation in patients with arterio-hepatic-venous shunts we performed embolization after temporary balloon occlusion of the hepatic vein through trans-jugular or transfemoral veins cannulation till the targeted hepatic vein and balloon were delivered in the shunt vein and inflated for 10–15 min during embolization time.
- In patients with portosystemic shunts a retrograde transcaval or transjugular obliteration of the shunt was performed under local anesthesia by advancing two catheters into the portal venous system. One catheter (Tracker 38, BSJ, Tokyo, Japan) was used and advanced into the main portal vein through the shunt to obtain a portogram and measure portal venous pressure before and during embolization and the other catheter was used to place the embolic material.

Different types of embolic agents were available and included fibered platinum coils, Gianturco steel coils, gel foam particles, polyvinyl ethaline particles (PVA, Ivalon Contour 350-500 emboli; Boston Scientific, Mississauga, ON, Canada), histoacryl (NBCA, *N*-butyl cyanoacrylate; B Braun, Melsungen, Germany) mixed with lipiodol (Laboratoire Guerbet,

Roissy, France) at a ratio of 1:2. The choice of embolic agent depended on the size and angioarchitecture of the shunt. In large simple or aneurismal shunts we used coil embolization. In complex shunts we used particle agents.

In patients with hepatic venovenous shunts due to underlying Budd-Chiari syndrome we performed TIPS (transjugular intrahepatic portosystemic shunt) using a non covered wall stent (Boston Scientific, Mississuauga, USA) between the IVC and intrahepatic portal vein main branches after balloon dilatation of the track.

2.5. Assessment of embolization results

We reviewed our patients who underwent shunt embolization and evaluated their radiological and clinical outcomes. The degree of shunt occlusion based on the pre- and post-embolization digital subtraction angiograms was evaluated and we divided it into two categories: complete and partial occlusion. Complete occlusion was defined as absence of residual opacification of the shunt and the draining veins and was considered immediate technical success. Partial occlusion was defined as visible evidence of a residual shunt and this was semi-quantitatively divided into two subcategories: those with $<80\%$ reduction and those with $\geqslant 80\%$ reduction.

Patients with 80% or more reduction were arranged for repeated shunt embolization 6 months afterward using more than one embolic material, if complete occlusion occured then no more treatment was needed. If failed, then surgery would be the next step. Patients with partial reduction of shunts $<80\%$ underwent direct surgery. We set the criteria at 80% reduction because the chance of sequential complete occlusion during follow-up would be high as small shunts can spontaneously resolve with conservative treatment as was previously reported [11].

3. Results

Among the 3143 patients included within the framework of screening, a total of 134 (4.3%) patients were finally diagnosed to have abnormal intrahepatic vascular shunts (IHVSs). They were 82 males and 52 females. Their ages ranged from 3 to 75 years (mean, 46 years). At the time of entry into the study, only 15 (11%) patients were presented with signs and symptoms specific for underlying shunts. The most common presentation was portal hypertension in 7 (5%) patients with arterioportal shunts. These patients had experienced various manifestations including splenomegaly in 7 (5%) patients, ascites in 6 (4.5%) patients, variceal hemorrhage in 6 (4.5%) patients and abdominal bruit in 2 (1.5%) patients (more than one presentation could be present in the same patient). Hepatic encephalopathy was manifested in 7 (5%) patients with portosystemic venous shunts in the form of disturbance of consciousness, tremors, disorientation, and somnolence. The most lethal presentation was high output heart failure in only one (1%) patient who had combined (arterioportal and arteriohepatic) shunt characterized by shortness of breath, dyspnea on exertion and lower limb edema. Eight patients (6%) with congenital arteriovenous shunts had associated extrahepatic manifestations of hereditary hemorrhagic telangiectasia (HHT) in the form of epistaxis and mucocutaneous telangiectases. Demographic and clinical characteristics are demonstrated in Table 1.

Table 1 Demographic and clinical characteristics of the diagnosed IHVSs in 134 patients.

Characteristics	Value data
Demographics	
Age (years)	
Mean (range)	46 (3–75)
Sex (number)	
Males	82
Females	52
Clinical presentation	No (%)
Portal Hypertension	7 (5%)
Hepatic Encephalopathy	7 (5%)
Heart Failure	1 (1%)
Extrahepatic HHT manifestations	8 (6%)

Two principle types of intrahepatic vascular shunts were finally diagnosed. Type I were arteriovenous shunts in 96 (72%) patients including isolated arterioportal shunts in 75 (56%) patients (Fig. 1), isolated arteriohepatic shunts in 20 (15%) patients (Fig. 2) and coexistence of both types of shunts in only 1 (1%) patient (fig. 3). Type II were venovenous shunts in 38 (28%) patients, including portohepatic venous shunts in 17 (13%) patients (Fig. 4), portoportal shunts in 14 (10%) patients (Fig. 5) and hepatic venovenous shunts in 7 (5%) patients (fig. 6).

The diagnosed intrahepatic vascular shunts (IHVSs) were mostly acquired due to underlying HCC which represented the most common etiology in 63 (47%) patients (Fig. 2). The size of their maximal individual tumors ranged from 4 to 10 cm in all patients. Acquired non neoplastic causes were determined in 63 (47%) patients including, transhepatic interventional procedures in 23 (17%) patients, liver cirrhosis in 14 (11%) patients (Fig. 4), traumatic liver injury in 11 (8%) patients, portal vein thrombosis in 10 (7%) patients (Fig. 6) and Budd-Chiari syndrome in 5 (4%) patients (Fig. 6). Congenital etiology of the shunts was determined in 8 (6%) patients with hereditary hemorrhagic telangiectasis (HHT) (Figs. 1 and 3). The different types of shunts and their associated etiologies are summarized in Table 2.

Imaging analysis of the size, angioarchitecture and distribution of shunts revealed that: 119 (89%) patients had small shunts (mean diameter, 4.5 mm, range 3–6 mm), 43 of them were simple and 76 were complex shunts [63 of them were associated with HCC including 48 arterioportal and 15 arteriohepatic (Fig. 2)]. Eleven (8%) patients had large shunts (mean diameter, 17 mm, range 15–23 mm) [10 were simple (fig. 4) and only one shunt was complex and diffusely distributed in the liver (fig. 3)] and 4 (3%) patients had aneurismal shunts (mean diameter, 32 mm, range 28–45 mm) (Fig. 1, Table 3).

Ultrasonography and color Doppler findings evaluation results differed according to the type, size, angioarchitecture and distribution of shunts. In small simple shunts ($n = 43$) there were no detectable morphological changes of the inflow and outflow vessels of the shunts and no alterations in their original flow velocity, pattern or direction as compared to the intrahepatic vessels of the same order.

- In small complex shunts ($n = 76$), the inflow and outflow vessels and their vascular connections appear as small reticular tortuous vascular network (Figs. 2A–C, 5A,B and

Table 2 Etiologies of the diagnosed IHVSs in 134 patients.

Etiologies of shunts	Types of shunts					
	Arteriovenous (96, 72%)			Venovenous (38, 28%)		
	Arterioportal (75, 56%)	Arteriohepatic (20, 15%)	Combined (1, 1%)	Portohepatic (17, 13%)	Portoportal (14, 10%)	Venovenous (7, 5%)
Acquired (126, 94%)						
Neoplastic						
HCC (63, 47%)	48	15	–	–	–	–
Non neoplastic						
Interventional procedures (23, 17%)	16	2	–	2	3	–
Cirrhosis (14, 11%)	1	–	–	13	–	–
Traumatic liver injury (11, 8%)	6	–	–	2	1	2
Portal vein thrombosis (10, 7%)	–	–	–		10	–
Budd Chiari syndrome (5, 4%)	–	–	–	–	–	5
Congenital						
HHT (8, 6%)	4	3	1	–	–	–

Table 3 Size and angioarchitecture of different types of IHVSs.

Type of shunts	Size and angioarchitecture of shunts				
	Small (119, 89%)		Large (11, 8%)		Aneurismal (4, 3%)
	Simple 43 (32%)	Complex 76 (57%)	Simple 10 (7%)	Complex 1 (1%)	
Arteriovenous (96, 72%)					
Arterioportal (75, 56%)	20	48	5	–	2
Arteriohepatic (20, 15%	5	15	–	–	–
Combined (1, 1%)	–	–	–	1[a]	–
Venovenous (38, 28%)					
Portohepatic (17, 13%)	10	–	5	–	2
Portoportal (14, 10%)	6	8	–	–	–
Hepatic venovenous (7, 5%)	2	5	–	–	–

[a] This shunt was diffusely distributed in the liver.

6B,C) and Doppler tracing revealed mild increase in flow velocities in arteriovenous shunts ($n = 63$) (mean systolic, 45; range 42–48 cm/s, and mean diastolic 18; range 17–22 cm/s), pulsatile biphasic waveform pattern with preserved flow direction in the draining portal and hepatic venous branches (Fig. 2E). In venovenous shunts ($n = 13$) the flow velocities inside the inflow and outflow vessels were decreased (mean, 10; range 9–15 cm/s, and mean, 22; range 20–26 cm/s in portoportal (Fig. 5C,D) and hepatic venovenous shunts (Fig. 6D,E), respectively), with continuous monophasic flow waveform patterns. The flow directions were reversed in the portal outflow vein (in portoportal shunts) and hepatic inflow vein (in hepatic venovenous shunts). The shunt ratios were $<30\%$ in all these patients (mean, 20%; range 11–25%).

- In patients with large ($n = 11$) and aneurismal ($n = 4$) shunts, blood was preferentially directed toward the shunt in the inflow vessels and away from the shunt in the outflow vessels in all the patients with consequent morphological changes and alterations in the Doppler spectra that differed according to the type of shunts as follows:

In patients with arteriovenous shunts (seven arterioportal and one combined) the feeding hepatic arteries showed increased diameters (mean, 66 mm, range 60–75 mm), tortuous courses, increased flow velocities (mean systolic, 100; range 80–120 cm/s, and mean diastolic 45; range 40–56) and decreased resistive indices (mean, 0.5, range, 0.45–0.6) associated with intrahepatic arterial hypervascularization. The draining veins showed increased diameters (mean, 13.5 mm, range 13.2–16.5 mm), increased flow velocities (mean 35 cm/s; range 27–60 cm/s) and arterialized biphasic waveform pattern. The flow inside the draining portal veins was reversed (Fig. 1B–F).

In patients with aneurismal portohepatic shunts ($n = 7$), the portal branches leading to the shunts were dilated (mean diameter, 15 mm,range 13–17 mm) with increased flow velocities inside (mena, 56; range 50–68 cm/s) associated with reduction of portal perfusion of the hepatic parenchyma as assessed by narrowing and slow flow in the intrahepatic portal branches not involved in the shunt. The draining hepatic veins showed increased diameters (mean, 13.3 mm, range 13–15 mm) associated with alteration in its Doppler spectra in the form high velocities (mean, 66; range 6–68 cm/s) and monophasic, continuous waveforms (Fig. 4B–F). Mean shunt ratio in all patients with large and aneurismal shunts was 55% (range 45–78%) (Table 4).

Images obtained from multiphasic CT protocol and post-processing techniques including MPR, MIP and VR allowed

Table 4 Ultrasonographic and color Doppler findings in the diagnosed 134 patients with IHVSs.

Ultrasonographic and color Doppler findings					
Size and appearance of shunts	Morphology of inflow and outflow vessels	Hemodynamic parameters			
		Velocities in inflow and outflow vessels	Waveform pattern in the outflow vessel	Flow direction in the outflow vein	Shunt ratio (mean, range)
Small					
Simple (n = 43)	No changes	No changes	No changes	No changes	20%; 11–25%
Complex (n = 76)	Reticular network	Mildly ↑ in AV ↓ in VV shunts	Altered in AV and VV shunts[a]	Preserved in AP Altered in VV shunts[b]	
Large (n = 11) and Aneurismal (n = 4)	Increased diameters and tortuous courses	*Arteriovenous shunts (n = 8)* – ↑ peak flow velocities and ↓ RI in the hepatic arteries – ↑ mean flow velocities in the portal veins	Arterialized, biphasic	Away from the shunt[c]	55%; 45–78%
		Portohepatic venous shunts (n = 7) – ↑ mean flow velocities in the portal veins – ↑ flow velocities in the draining hepatic veins	Continuous, monophasic	No change	

Abbreviations: AV = arteriovenous shunts, VV = venovenous shunts.
[a] Pulsatile biphasic in arteriovenous shunts and continuous monophasic in venovenous shunts.
[b] Reversed flow was observed in the outflow vein in portoportal shunts and in the inflow vein in hepatic VV shunts.
[c] Inversion of flow direction was observed in the draining portal veins.

us to directly visualize the communications between inflow and outflow vessels in all projections. The type, size, angioarchitecture and distribution of shunts as well as the identity, diameters and courses of the inflow and outflow vessels were confirmed by CT. Other additional indirect CT signs allowed us to identify and differentiate between various types of shunts. In all patients with arteriovenous (arterioportal and arteriohepatic) shunts (n = 96) we observed simultaneous opacification of the hepatic artery (inflow vessel) and the outflow venous branches (portal vein in arterioportal shunts (Fig. 1G–M, hepatic veins in arteriohepatic shunts (Fig. 2F,G) or both veins in combined shunts (Fig. 3E–G)) during the early arterial phase. This early venous enhancement had approximately the same attenuation as that of the hepatic arteries and persisted during the late arterial and venous phases. In patients with portovenous shunts (n = 38) the portal and hepatic veins did not opacify in the arterial phase but both of them opacified simultaneously during the porto-venous phase (Fig. 4G–L).

Intraparenchymal perfusion disorders [transient hepatic attenuation differences (THADs)] were another indirect signs observed in a total of 16 (12%) of patients with IHVS; 10 patients had arterioportal shunts, 1 patient had combined arterioportal and arteriohepatic shunts associated with HHT (parenchymal hepatic telangiectasis) (Fig. 3 E–G) and the remaining 5 patients had obstructed hepatic veins due to Budd-Chiari syndrome (Fig. 6 F,G). These perfusion disorders appeared as peripheral wedge or globular shaped areas with regular contours characterized by early enhancement that persisted during both arterial phases and either returned to normal attenuation on portal-venous phase images in arteriovenous (AV) shunts or persisted in the venous phase in venovenous (VV) shunts due to venous occlusion. The edge of the wedge shaped areas of THADs, pointed toward the portal hilus in arterioportal shunts and toward the vena cava in hepatic vein occlusions. The intrahepatic CT findings of shunts are summarized in Table 5.

Table 5 CT findings in the diagnosed 134 patients with IHVSs.

	CT findings	
	Arteriovenous shunts (n = 96)	Venovenous shunts (n = 38)
I. Time of opacification of shunt vessels		
Early arterial phase	–Simultaneous opacification	–No opacification
Late arterial phase	–Persistent opacification	–No opacification
Portovenous phase	–Persistent opacification	–Simultaneous opacification
II. Parenchymal perfusion disorders (n = 16)		
–Shape and distribution	–Peripheral wedge shaped or globular areas	
–Time of opacification		
• Early arterial phase	–Early enhancement	
• Late arterial phase	–Persistent enhancement	
• Portovenous phase	–Isodense to the liver in AV shunts or persisted enhancement in VV shunts	
–Direction of the wedge of THAD	Arteriovenous shunts (n = 11)	Venovenous shunts (n = 5)
	Pointed toward the portal hilus	Pointed toward the vena cava

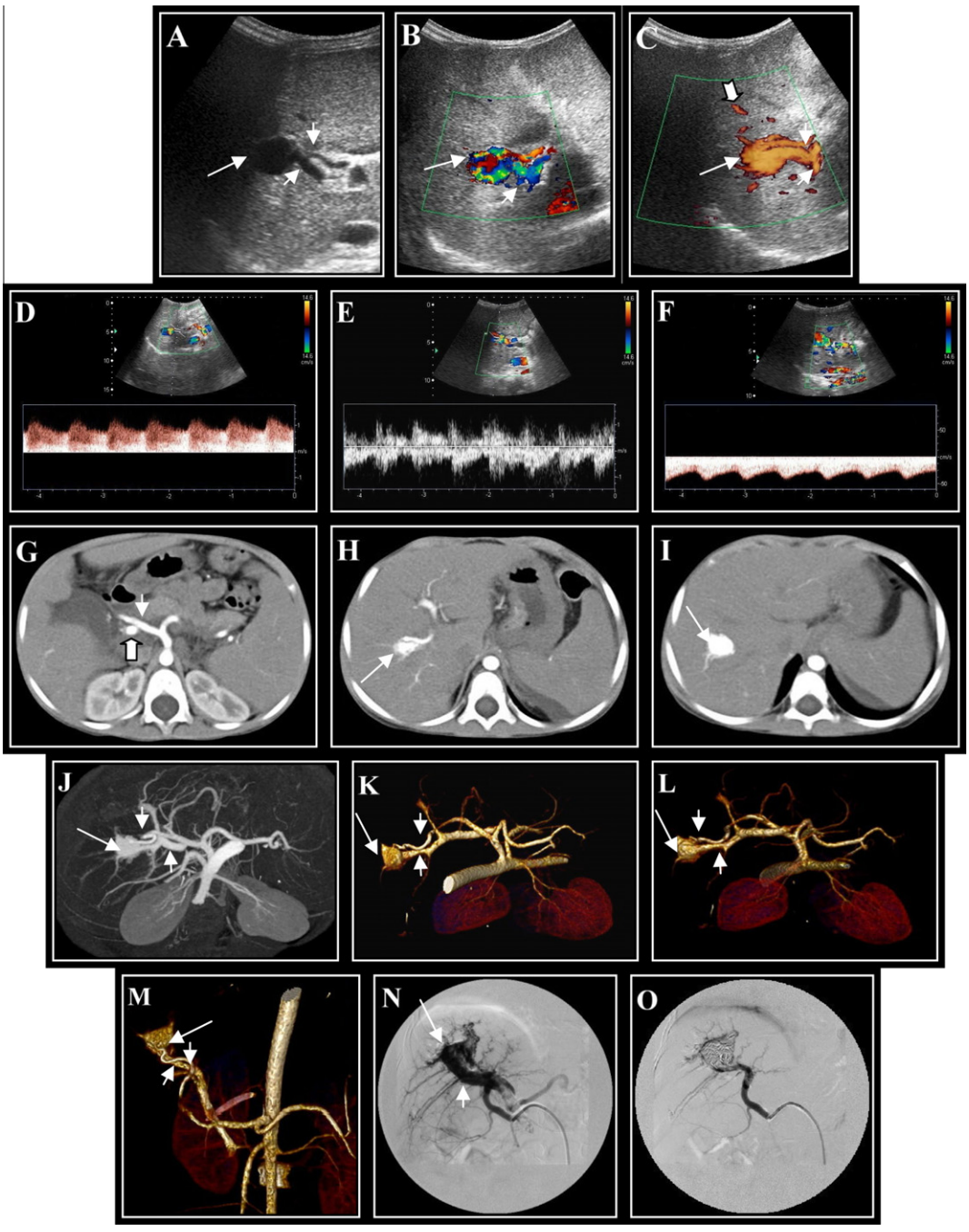

Figure 1 A 5-year-old child with congenital aneurismal arterioportal shunt, manifested by portal hypertension. (A) Gray-scale sonogram shows a cystic structure (long arrow) adjacent to the hepatic vessels which are dilated and tortuous (short arrows). (B) Color Doppler ultrasound clearly identifies direct connection of the hepatic artery to the portal vein via a vascular aneurysm (long arrow) with reversal of flow in the portal vein (short arrow). (C) Power Doppler sonogram shows adequate morphological evaluation of the aneurismal shunt (long arrow), feeding and draining vessels (short arrows) and associated hypervascularity of the surrounding liver parenchyma (thick arrow). (D–F) Spectral Doppler tracing of the hepatic artery shows high systolic and diastolic flow velocities (D), bidirectional flow throughout the aneurismal shunt (E) and arterialized reversed flow in the draining portal vein (F) consistent with arterioportal shunting. (G–I) Early arterial phase axial CT scans at different levels show early opacification of the portal vein (thick arrow) adjacent to the hepatic artery (thin arrows) (G), confirming arterioportal shunt. The aneurismal site of communication is seen on cranial CT scans (H and I) (arrows). Three dimensional MIP (J) and VR angiographic reconstructions (K–M) allow better angiographic details of the aneurismal shunt (long arrows), inflow and outflow vessels (short arrows) in axial anterior (K), posterior (L) and anterior coronal (M) orientations. (N) Pre-embolization selective DSA of the main hepatic artery confirms the aneurismal arterioportal shunt (long arrow) with retrograde filling of the main portal vein (short arrow). (O) Postembolization assessment revealed complete occlusion of the shunt by coils.

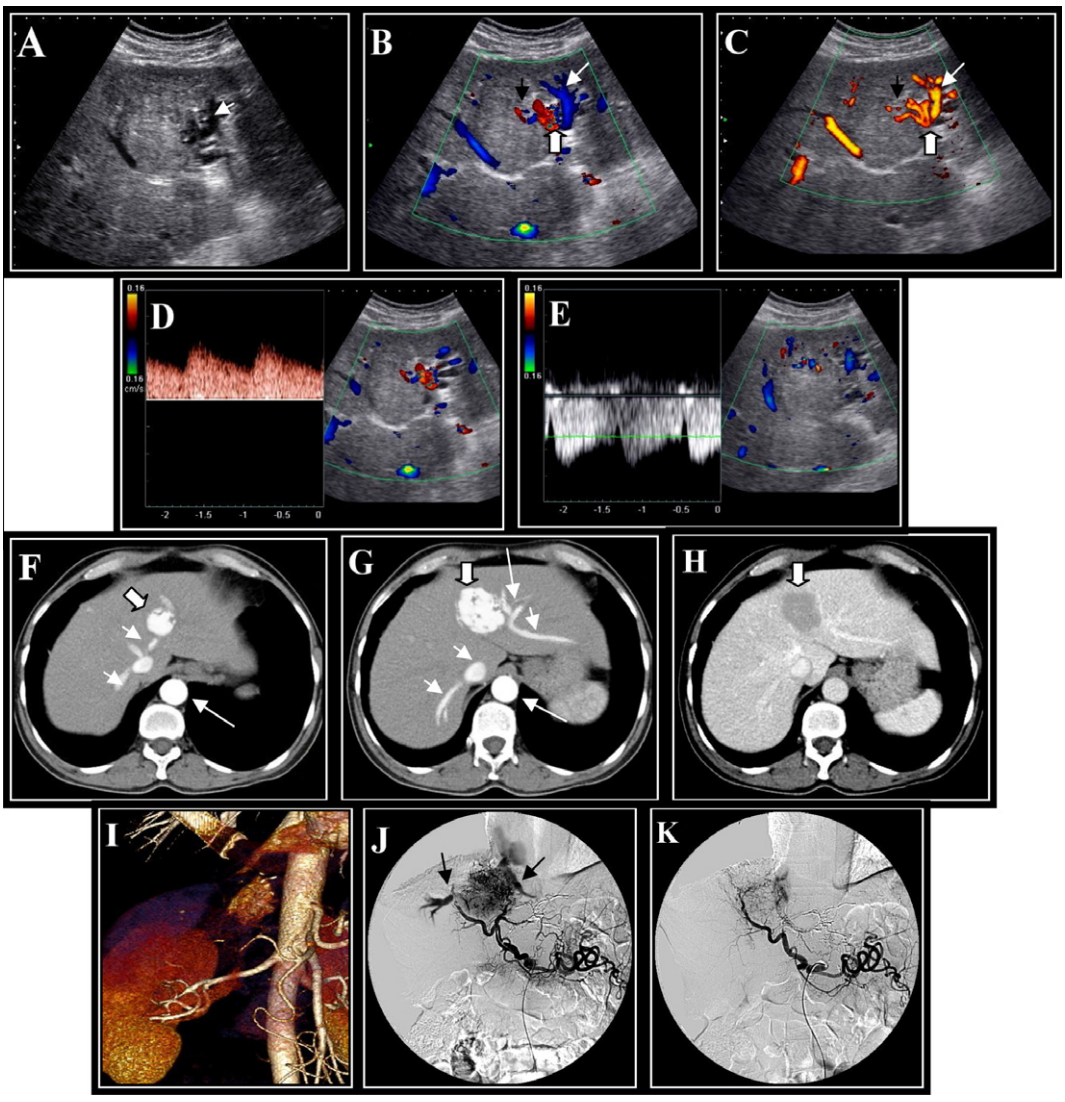

Figure 2 A 55-year-old woman with hepatocellular carcinoma and arteriohepatic venous shunt. (A) Gray-scale sonogram shows an isoechoic mass in the left lobe of the liver surrounded by anechoic tortuous and tubular structures (arrow). (B) Color flow mapping, and (C) power Doppler images show a complex shunt (thick arrows) communicating intra- (black arrows) and perilesional (white arrows) vessels. (D and E) Spectral Doppler tracing reveals mild increased systolic and diastolic velocities in the intralesional arterial vascularity (D) with pulsatile biphasic flow in the adjacent hepatic vein (E) consistent with arteriohepatic venous shunting. (F and G) Contrast-enhanced axial CT images in the early arterial phase at two levels show simultaneous filling of contrast of the aorta and hepatic arteries (long arrows) and hepatic veins and IVC (short arrows) representing arteriosystemic venous shunting due to a heterogeneously enhanced mass (thick arrows) that shows early venous drainage with complete washout of its contrast in the venous phase image (thick arrow) (H) consistent with HCC. (I) Three dimensional oblique coronal VR angiographic reconstruction image allows good morphological and angiographic details. (J) Pre-embolization selective DSA of the main hepatic artery confirms the presence of neoplastic arteriosystemic shunt with retrograde filling of the draining hepatic veins adjacent to the mass (black arrows). (K) Postembolization assessment revealed $<80\%$ reduction of the shunt and the patient underwent surgery.

Following the protocol of management proposed by our team we found that: 56 (42%) asymptomatic patients had small non neoplastic shunts and their shunt ratios were $<30\%$ (group I). These patients were subjected to close monitoring by duplex Doppler and clinical follow-up. After 3 months regression of the shunts was observed in 33 patients (20 had arterioportal and 5 had arteriohepatic venous shunts and 8 portoportal shunts), while in 18 patients (10 had portohepatic 6 had portoportal and 2 had hepatic venovenous shunts) the shunt sizes remained constant, and all these patients did not develop any signs or symptoms and their shunt ratios were $<30\%$. In the remaining 5 patients with hepatic venovenous shunts, new shunts developed, shunt ratios exceeded 30%, with marked attenuation of the hepatic veins after 6–12 months and the patients developed manifestations of complications (ascites and varices). These patients underwent TIPS.

Seventy-seven (57%) patients were categorized under group II. This group included 14 symptomatic patients with shunt ratios exceeded 40%; 10 patients had large simple shunts and 4 pa-

tients had aneurismal shunts. Another 63 patients who had small complex shunts associated with HCC were also categorized under this group. Interventional treatment was immediately applied for these patients including coil embolisation in patients with large or aneurismal shunts and chemoembolization using histoacryl or gel foam with simultaneous injection of chemotherapeutic agent (TACE) in patients with HCC. Postembolization assessment revealed complete occlusion in a total of 12 (8%) patients (10 had large simple shunts and 2 had aneurismal shunts), >80% reduction in a total of 56 (42%) patients (2 had aneurismal shunts and in 54 had neoplastic shunts) and <80% reduction in the remaining 9 (7%) neoplastic shunts. Subsequent embolization was arranged 6 months afterward for the 56 patients with >80% reduction and we could no longer monitor any shunts in postembolization angiograms. Patients with <80% reduction of their neoplastic

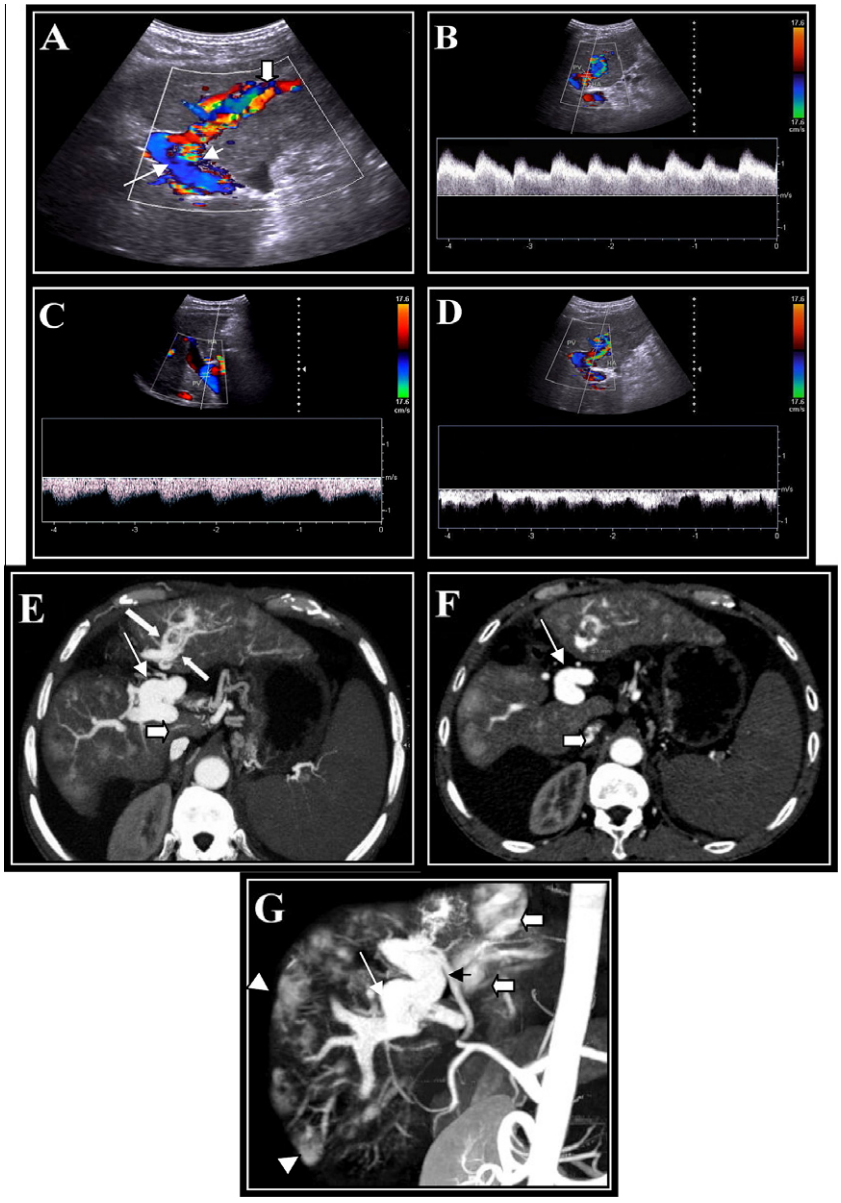

Figure 3 A 6-year-old child with HHT, combined arterioportal and arteriosystemic venous shunts and hepatic perfusion disorders manifested by heart failure. (A) Color flow mapping shows dilated tortuous portal vein (long arrow), and hepatic artery (short arrow) associated with a vascular network inside the left liver lobe (thick arrow). (B–D) Spectral tracing reveals involvement of the hepatic artery, portal vein and hepatic vein branches in this network with high systolic and diastolic flow velocities in the hepatic artery (B), and arterialized flow in the portal (C) and hepatic (D) veins with reversed flow in the portal vein consistent with a combined arterioportal and arteriosystemic shunt. (E and F) Axial CT images at two different levels and (G) coronal oblique 3D MIP image during the early arterial phase show simultaneous opacification of the hepatic veins and IVC (thick arrows), portal vein (long arrows), and hepatic artery (black arrow), better evident in the coronal image (G) due to underlying combined arteriosystemic and arterioportal intraparenchymal shunts (between arrows). The liver parenchyma is heterogeneous because of the presence of several telangiectases with multiple peripheral triangular and globular enhancing areas (THADs) (arrowheads) due to underlying parenchymal perfusion disorders.

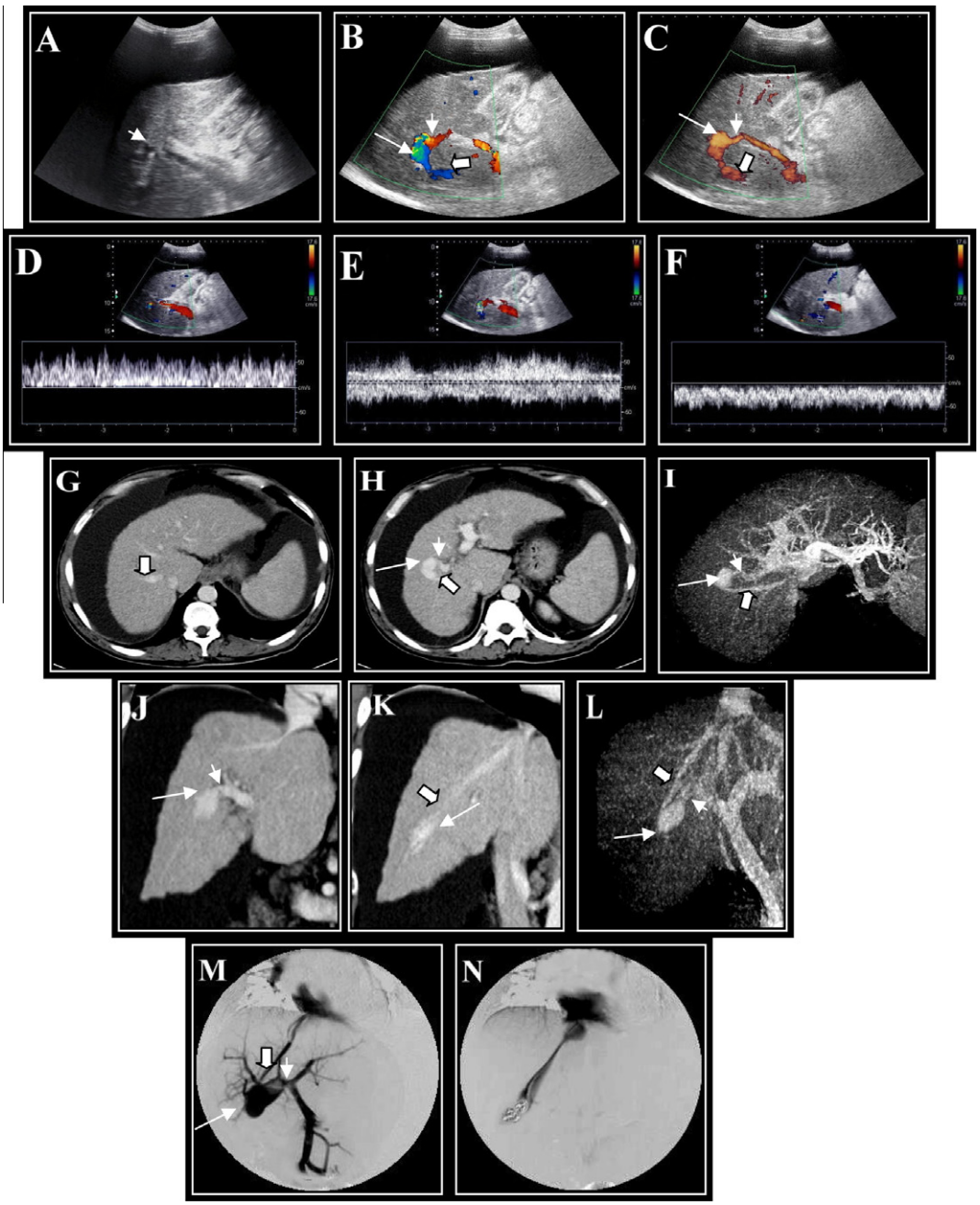

Figure 4 A 60-year-old male patient with aneurismal portohepatic venous shunt, undergoing evaluation for cirrhosis and presented by hepatic encephalopathy. (A) Gray scale sonogram shows a cystic appearing lesion adjacent to the hepatic vessels (arrow). Color flow (B) and power Doppler (C) sonograms show direct connection of the right portal vein branch (short arrows) with the right hepatic vein (thick arrows) through an aneurysm (long arrows). (D–F) Doppler spectral tracing shows a high flow velocity with cardiac modulation of the spectrum within the portal vein due to the transmission through the shunt of the cardiac related flow oscillations of the efferent hepatic vein (D), helicoidal bi-directional wave within the aneurismal portovenous shunt (E), and a flattened, turbulent, high velocity waveform within the draining hepatic vein (F). Axial (G–I) and coronal (J–L) reformatted contrast-enhanced portal venous phase and angiographic MIP CT images show an aneurismal shunt communication (long arrows) between the right portal vein (short arrows) and the right hepatic vein (thick arrows). (M) Pre-embolozation hepatic venogram shows intrahepatic portosystemic venous shunt (long arrow) between the right portal vein (short arrow) and the right hepatic vein (thick arrow). (N) Postembolization assessment shows complete obliteration of the shunt by coils.

shunts proceeded to surgical reconstruction of shunts and hepatectomy after satisfactory shunt and tumor control by chemoembolization.

Group III included only one (1%) patient with combined (arterioportal and arteriohepatic) complex shunt diffusely distributed in the liver. This patient had shunt ratio $>60\%$ and

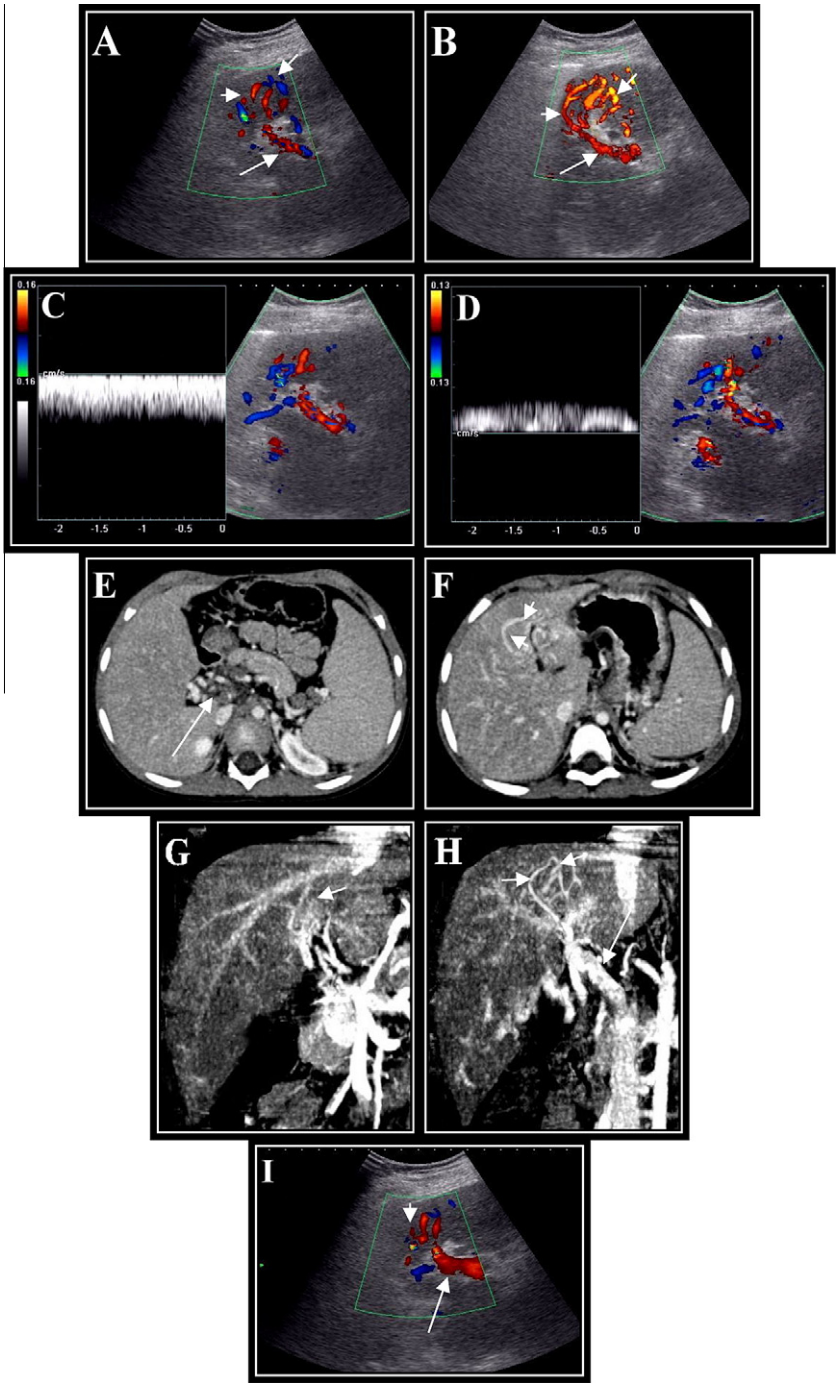

Figure 5 A 45-year-old male patient with portoportal shunts due to underlying main portal vein thrombosis. (A and B) Color and power Doppler sonograms show cavernous transformation of the thrombosed main portal vein (long arrows) with a reticular network of small portal vessels at and close to the liver hilum connecting the left portal branches with the right portal branches (short arrows). (C and D) Spectral tracing shows slow flow velocities with continuous monophasic flow pattern in the inflow and outflow vessels and reversed flow in the outflow vessels. Portovenous phase axial (E and F) and coronal MIP (G and H) images confirm the ultrasonographic finding of cavernous transformation of the thrombosed main portal vein (long arrows) with direct portoportal connections (short arrows). (J) Follow-up color flow mapping shows partial recanalization of the main portal vein (long arrow) with regression of the size and number of the shunt vessels (short arrow).

was presented by heart failure. He directly underwent liver transplantation.

Close clinical and ultrasonographic follow-up was applied and advised at 6–12 months intervals for all the patients and prophylactic medical therapy which consists of a protein-free diet and oral administration of lactulose, branched-chain amino acids and non absorbable antibiotics was administered for patients with portovenous shunts and anticoagulant therapy

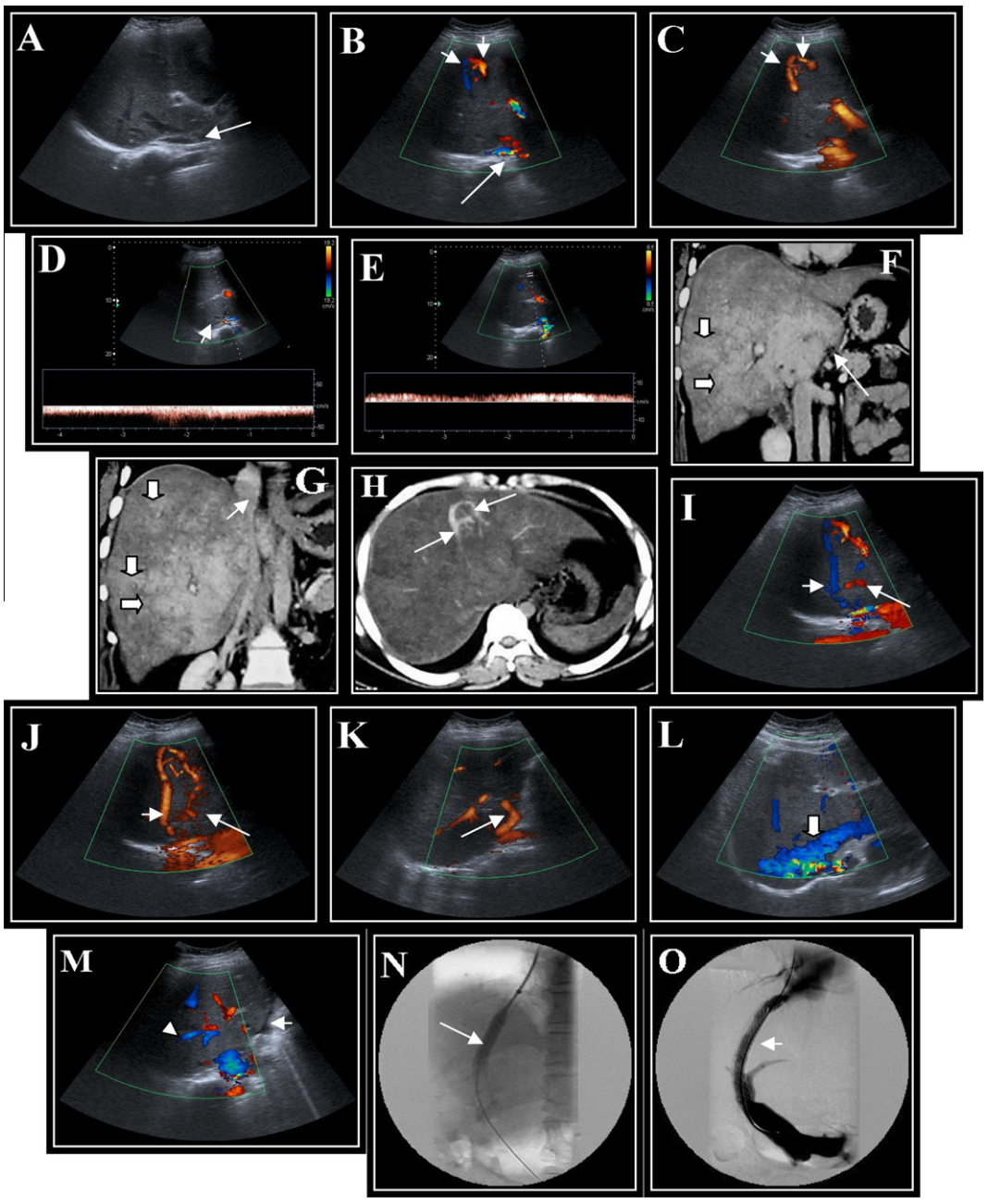

Figure 6 A 21-year-old female patient with hepatic venovenous shunts due to underlying Budd-Chiari syndrome. (A) Gray scale US demonstrates an echogenic web constricting the IVC (long arrow). (B and C) Color and power Doppler sonograms show focal color aliasing in the IVC (long arrow) indicating a stenosis at that level and a network of fine venovenous collateral vessels (short arrows). (D and E) Doppler tracing of the IVC (D) and collateral vessels (E) show weak monophasic continuous flow with loss of the normal triphasic appearance. Note and a thrombus narrows hepatic vein confluence (short arrow). (F and G) Coronal portovenous phase images show inhomogeneous enhancement of the liver, enlarged caudate lobe (long arrow) with wedge shaped and globular areas of hepatic attenuation differences (thick arrows) persisted during the portovenous phase with partial obstruction of the IVC by a thrombus (short arrow). Axial MIP (H) image obtained from portovenous phase confirm the presence of spider web network of venovenous collaterals (arrows). (I–M) Color and power Doppler follow up after 6 months shows development of new collaterals (short arrows) that follow the same track of the stenosed right hepatic vein and connecting it to neighboring middle hepatic vein results in presence of ''bicolored'' hepatic veins (long arrows). A large caudate lobe vein (K) (long arrow) and a large tortuous subcapsular vein (L) (thick arrow) that drain directly into the IVC were developed and reversed flow was observed in the main and branches of the right portal vein creating new intrahepatic and extrahepatic shunts (M) (arrowhead). Note the presence of ascites (short arrow). The patients presented by manifestations of portal hypertension. TIPS was performed with balloon dilatation of the track through a transjugular approach (N) (long arrow) then a stent was placed to connect the IVC with the portal vein (O) (short arrow).

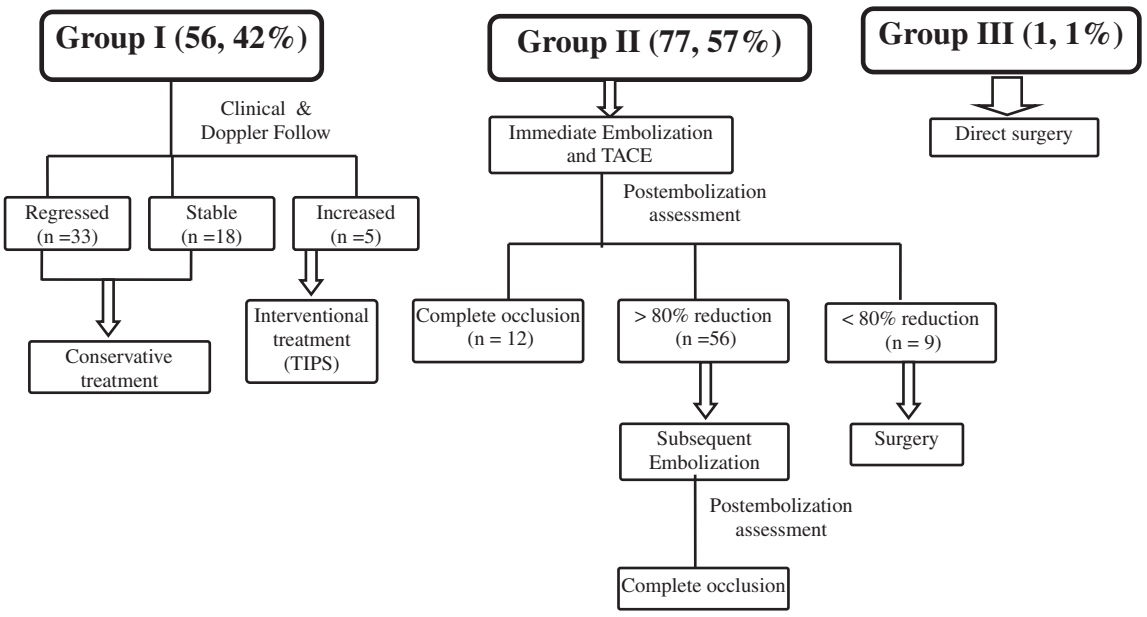

Figure 7 Chart illustrates application of the proposed strategy for managing the diagnosed 134 patients with IHVSs.

was administered for patients with hepatic venovenous shunts to prevent further development of complications. The algorithm used for management is illustrated in Fig. 7.

4. Discussion

Diagnosis of intrahepatic vascular shunts (IHVSs) has been long considered challenging and many times it is made incidentally [1,10,12]. Early diagnosis and proper management of intrahepatic vascular shunts is very important in both symptomatic and asymptomatic patients [12]. Our study findings confirmed what was previously reported by many investigators (12-17) as we found that liver involvement by intrahepatic vascular shunts was asymptomatic in the majority 89% of our patients, and when symptomatic (in 11%) they were presented by severe complications including portal hypertension (in 5%), portosystemic encephalopathy (in 5%), and congestive cardiac failure (in 1%). Moreover, recent publications [13–15] reported that the interval between the formation of the fistula and its recognition can range from hours to years. That is why we screened a large group of patients who were liable to develop intrahepatic vascular shunts aiming to reduce the risk of deterioration of serious complications, or to prevent their development. Due to its ease of use, non-invasiveness and availability, we proposed ultrasonography and color Doppler imaging as the initial screening test. We complemented our imaging evaluation of the detected IHVSs by a more accurate imaging method using multidetector CT protocols as detection of these malformations should always prompt careful evaluation in an attempt to identify other diagnostic criteria and confirm ultrasonographic findings. Angiography is an invasive and expensive procedure that previously was the most useful imaging method for the assessment of vascular abnormalities [12,16,17], but in our study, considering the fact that liver involvement is asymptomatic in most cases, it was indicated only in selected cases for interventional management (Figs. 1,2,4,6).

Previous publications [1,2,4,5,8,11] had reported that three types of intrahepatic shunts between the major vessels of the liver were possible including: arterioportal shunts (hepatic artery to portal vein), arteriosystemic shunts (hepatic artery to hepatic vein), and portosystemic venous shunts (portal vein to hepatic vein or vena cava). Bodner et al. [6] added another two types to their classification including portoportal shunts (between portal veins) and hepatic venovenous shunts (between hepatic veins). In order to simplify their description we classified the diagnosed IHVSs in 134 patients into two major types; arteriovenous shunts in 72% and venovenous shunts in 28% of our patients. We found that arterioportal shunts, were the most common type identified in 57% of our patients. They were presented as isolated arterioportal shunts in 56% of patients and coexisted with arteriohepatic shunts in only 1% of our patients. Portohepatic shunts were identified in 13%, portoportal shunts in 10% and hepatic venovenous shunts in 5% of our patients.

Our study results have proved that intrahepatic vascular shunts were more frequent than was previously reported by other authors [18–33]. Intrahepatic vascular shunts were identified in 4.3% of the screened 3143 patients. This is likely as we relied mainly on imaging findings for diagnosis as compared with others [26,28,31] who depended on clinical findings alone and also due to the higher general sensitivity of the imaging methods we used in detecting hepatic alterations. This is also attributed to the prevalence of intrahepatic vascular shunt particularly arterioportal and arteriosytemic shunts in patients with HCC (47%) and in patients who underwent percutaneous transhepatic procedures (17%) and moreover, due to the high incidence of portosystemic shunts in patients with cirrhosis (11%). These patients constituted a large group of patients who were routinely managed in our departments. Researchers show that the prevalence of arterioportal shunting in HCC is as high as 63% of patients [1,12,13,17]. Other articles stated that arteriosystemic shunts are typically associated with benign and malignant liver neoplasms. This was explained by the fact

that the hepatic artery supplies HCC almost exclusively and the tendency for these neoplasms to grow in the portal or hepatic veins causes the venules to act as efferent vessels for the tumor [1,6,7]. A previous article had reported that incidence of intrahepatic shunts has increased in the last years because of the greater use of transhepatic interventional procedures [1]. In our study the prevalence of portosystemic shunts in patients with liver cirrhosis, portoportal shunts in patients with portal vein thrombosis and hepatic venovenous shunts in patients with Budd-Chiari syndrome were nearly the same as the prevalences previously reported by many investigators [6,32,34,35]. We found that congenital etiology due to underlying hereditary hemorrhagic telangiectasis was determined in 6% of our diagnosed shunts and were mainly associated with arteriovenous shunts.

Diagnostic imaging has a fundamental role in the identification of hepatic vascular alterations. Apart from diagnosis and differentiation of various types of shunts it is crucial to evaluate the shunt comprehensively in order to select appropriate management specific for each patient [12,17]. Color Doppler ultrasonography (CDUS) has proved useful in the detection of various intrahepatic vascular fistulas and has the additional advantage over other imaging modalities that it enables flow direction, flow velocity, and type of blood flow (i.e., arterial, portal, or hepatic venous) to be determined noninvasively [2,3,6,9,10]. These facts had made color Doppler ultrasonography very beneficial for imaging evaluation of IHVSs in our study as it provided us with baseline qualitative and quantitative morphological and hemodynamic road maps of the liver vasculature that helped us for selection of the appropriate management and follow up purposes. Recent publications concluded that it obviates the need for angiography as it is also able to measure shunt ratios and may be potentially useful in follow up and helping choose the appropriate therapeutic option of IHVS [2,3,10], although the main limitation is low sensitivity and spatial resolution in the detection of small arteriovenous shunts [2,3]. In our study we did not meet this limitation as we did not rely for diagnosis of small shunts (89%) on direct visualization of the shunt only but we also we routinely use spectral analysis of shunt vessels for all the patients and confirm our evaluation by multiphasic CT. In addition, ultrasonography combined with color Doppler allowed us to diagnose all the patients with large and aneurismal shunts (11%) easily and confirmed the previously reported sonographic criteria for liver involvement by shunts which included dilatation and increased flow velocity of the involved vessels associated with inversion and arterialization of the draining portal flow in arterioportal shunts and disruption of the normal triphasic pattern of the draining hepatic vein in arteriohepatic shunts [2–6]. The diagnosis of intraparenchymal portosystemic fistulas is based on direct visualization of a dilated portal vein communicating directly with a hepatic vein [21,25,26]. The high accuracy of these criteria for detecting intrahepatic shunts has led some authors to consider CDUS sufficient for diagnosis in asymptomatic subjects, with more invasive tests, such as CT, MRI or DSA being reserved for symptomatic subjects [36–38].

The introduction of multidetector row CT (MDCT) devices which allow for a dynamic multiphase study has facilitated detection of IHVS. The examination protocol consisting of early and delayed arterial phases and a portovenous phase and significant findings for identifying and differentiating the various types of shunts have been standardized [1,39–41]. Using multidetector row CT scanners in our study, enabled us to perform a complete multiphasic study of the hepatic vascular system and a good quality multiplanar reformatting (MPR) and angiographic (maximum intensity projection, MIP; and volume rendering, VR) reconstructions, thus offers a more accurate and global visualization of these shunts. In addition we relied on other indirect findings based on the time of opacification of the affected vascular branches during multiphasic study as well as the underlying parenchymal perfusion changes that could not only diagnose the presence of shunt but also specify the type of shunt and the vessels involved. Intrahepatic perfusion disorders, in the form of transient hepatic parenchymal enhancement were observed in 16 of our patients and were particularly associated with arterioportal shunts in 10 patients. These enhancement disorders were also observed in I patient with combined arterioportal and arteriohepatic shunts and in 5 patients with Budd-Chiari syndrome. They were presented typically as was previously described [37,39] as wedge-shaped or globular areas of enhancement during the arterial phase, which become isoattenuating in the portal venous phase in arteriovenous shunts but persisted in patients with hepatic vein obstruction.

The optimal therapy for IHVS remains controversial. Definitive therapy aimed at obliteration of the shunt and restoration of normal hemodynamics of the liver vasculature [12,42]. Herein, we proposed a practical strategy based on our experience and review of the literature that categorized patients into three groups according to the line of management that was considered suitable according to the clinical and imaging findings of each group. The specific way of management we selected depended on whether the patient was symptomatic or not, and also on the size of the shunt, shunt ratio and whether it was associated with HCC. Previous publications had reported that large intrahepatic shunts are more often associated with clinical manifestations of shunt complications than small shunts, as the higher degree of shunting due to larger shunts were responsible for early clinical manifestations. When the shunt ratio is less than 30% symptoms associated with IHVS may not develop throughout the life time of the individual. When the shunt ratio exceeds 30%, clinical manifestations may develop at any time. When the shunt ratio exceeds 60% even without manifestations the risk of complications is increased this is an indication for immediate intervention [2,4,9,12].

Emergency intervention was not required in 56 (42%) of our patients categorized under group I. These patients had small non neoplastic shunts and their shunt ratios were $< 30\%$ and consequently the patients were asymptomatic and there was enough time to follow-up their shunts and re-evaluated them, both clinically and by duplex Doppler examination. As estimated in previous articles [12,17,34] small shunts thrombosed and closed spontaneously in most patients. In our study, the shunts regressed in 33 patients, while remained unchanged in 18 patients after 3 months by follow-up duplex Doppler examinations. However, in the remaining 5 patients who had Budd-Chiari syndrome and hepatic venovenous collaterals new shunts developed and the patients had manifestations of complications and underwent TIPS. Embolization was clearly indicated in a total of 77 (57%) patients constituting group II to avoid more deterioration in 14 symptomatic patients with large and aneurismal with shunt ratio exceeding 40% or to improve

the prognosis in 63 patients who had HCC with rather small, complex shunts. Our concept was to treat the shunt and HCC at the same setting. Transcatheter embolization now affords a relatively safe, inexpensive and practical method for treatment of IHVS [25,32,43–45]. Compared with surgery, it carries the advantage of reduced morbidity, repeated access availability and reduced costs. Shorter hospital stay and decreased pain are specific advantages [1,43,44]. Different embolic agents were reported to achieve shunt embolisation with different degrees of occlusion [12,19,25,32,43–45].

The reason for choosing particular embolic agents is seldom mentioned [7]. In the current study, our choice of embolic agents was based on good understanding of the underlying mechanism of the shunts and their angioarchitecture. We preferred to use coils in 14 patients with large simple and aneurismal shunts than gel foam particles, because the latter could have been passed through the fistula and ended up in the splanchnic circulation. Embolization alone using coils achieved complete occlusion of the shunts with subsequent disappearance of symptoms in 12 patients, and $>80\%$ reduction of shunts with improvement of the patients' clinical condition in another 56 patients (2 had aneurismal and 54 had neoplastic shunts). This could be due to the use of relatively small coils in the 2 patients with aneurismal shunts or the complex nature of neoplastic shunts. Repeated embolization was successfully performed for them 6 months later using larger coils in aneurismal shunts and histoacryl combined with gel foam in neoplastic shunts. We yet achieved $<80\%$ reduction of shunts in 9 patients with HCCs but at least the chemoembolization procedures downgraded the HCCs and improved the prognosis for these patients. These patients needed further surgery as the next step and surgical approaches included vascular repair of the shunt and partial hepatectomy.

Liver transplantation was the only suitable approach selected for one of our patients who had congenital coexistence of arterioportal and arteriosystemic shunts. The shunt was diffusely distributed in the liver and we expected great difficulty to be managed by embolization particularly with the very bad clinical manifestation of the patient. To date, no cases of congenital shunts closed spontaneously have been reported. In such cases with advanced symptoms, the therapeutic role of hepatic arterial embolization has been proposed [28,30] but is controversial because of its unpredictable results and association with a high risk of fatal hepatic necrosis; thus, liver transplantation remains the only therapeutic possibility [4,5,12].

5. Conclusion

Routine follow-up of patients who were liable to develop intrahepatic vascular shunts using color duplex Doppler ultrasonography complemented by multiphasic multidetector CT protocols and familiarity of the imaging characteristics of various types of shunts allowed early and accurate diagnosis and comprehensive evaluation of intrahepatic vascular shunts. This was important to prevent development of serious complications or more deterioration of the affected patients. This was achieved by early selection of the appropriate line of management suitable for each patient. Based on combined imaging and clinical findings, our management strategy allowed uniform and practical therapeutic recommendations and yielded satisfactory results and good prognosis for all patients.

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